

IN THE CLAIMS

1. (currently amended) A medical system for treating spinocerebellar ataxia type 1 in a human patient comprising:

(a) an intracranial access device;

(b) a ~~patient-specific intraoperative~~ mapping means for locating a predetermined location in the brain of the patient, said location comprising cells natively expressing a gene encoding an ataxin-1 protein;

(c) a deliverable amount of a small interfering RNA capable of reducing the amount of ataxin-1 protein produced in said cells or a vector encoding said small interfering RNA wherein said small interfering RNA has a length of between about 15 and about 30 nucleotides; and

(d) a delivery means for delivering said small interfering RNA or vector encoding said small interfering RNA to said location of the brain of said patient from said intracranial access device through a stereotactically implanted catheter, said catheter comprising a radiographic marker material which does not interfere with intra-operative brain imaging, wherein said small interfering RNA hybridizes to a sequence identical to SEQ ID NO: 1 within the ataxin-1 mRNA.

2–9. (Cancelled)

10. (original) A medical system of claim 1 wherein said intracranial access device is an intracranial access port.

11-13. (Cancelled)

14. (original) A medical system of claim 1 wherein said predetermined location is the dentate nucleus, emboliform nucleus, the globose nucleus, the fastigial nucleus of the cerebellum (collectively the deep cerebellar nuclei), or the cerebellar cortex.

15-23 (Cancelled)

24. (previously presented) A medical system of claim 1 wherein said intracranial access device comprises an intracranial access port and wherein said delivery means is injection from an external syringe into the intracranial access port .

25. (original) A medical system of claim 1 wherein said delivery means is an infusion pump.

26-85 (Cancelled)

86. (previously presented) A medical system of claim 1 wherein said small interfering RNA inhibits the translation of ataxin-1 protein.

87. (Currently Amended) A medical system of claim 1 wherein said small interfering RNA reduces levels of mRNA encoding ~~for~~ ataxin-1 protein.

88. (Canceled).

89. (previously presented) A medical system of claim 1 wherein said small interfering RNA has sufficient complementarity to the ataxin-1 mRNA for said small interfering RNA molecule to direct cleavage of said ataxin-1 mRNA via RNA interference.

90. (Currently Amended) A medical system for treating spinocerebellar ataxia type 1 in a human patient comprising:

(a) an intracranial access device comprising a radiographic marker ~~material that does not interfere with intra-operative brain imaging;~~

(b) a ~~patient specific intra-operative~~ mapping means for stereotactically locating a predetermined location in the brain of the patient, said location comprising cells natively expressing a gene encoding an ataxin-1 protein;

(c) a deliverable amount of a small interfering RNA capable of reducing the amount of ataxin-1 protein produced in said cells or a vector encoding said small interfering RNA, said small interfering RNA comprising any one of SEQ ID Nos: 1 and 2; and

(d) a delivery means for delivering said small interfering RNA or vector encoding said small interfering RNA to said location of the brain of said patient from said intracranial access device through a stereotactically implanted catheter.

91. (previously presented) A medical system of claim 90 wherein said intracranial access device is an intracranial access port.

92. (previously presented) A medical system of claim 90 wherein said predetermined location is the dentate nucleus, emboliform nucleus, the globose nucleus, the fastigial nucleus of the cerebellum (collectively the deep cerebellar nuclei), or the cerebellar cortex.
93. (previously presented) A medical system of claim 90 wherein said intracranial access device comprises an intracranial access port and wherein said delivery means is injection from an external syringe into the intracranial access port .
94. (previously presented) A medical system of claim 90 wherein said delivery means is an infusion pump.
95. (previously presented) A medical system of claim 90 wherein said small interfering RNA inhibits the translation of ataxin-1 protein.
96. previously presented) A medical system of claim 90 wherein said small interfering RNA reduces levels of mRNA encoding for ataxin-1 protein.
97. (previously presented) A medical system of claim 90 wherein said small interfering RNA has sufficient complementarity to the ataxin-1 mRNA for said small interfering RNA molecule to direct cleavage of said ataxin-1 mRNA via RNA interference.
98. (Currently Amended) A medical system of claim 1 wherein said small interfering RNA comprises ~~any one of~~ SEQ ID Nos: ~~1 and 2~~ and a sequence at least 90% complementary thereto.